Management of cervical ribs and anomalous first ribs causing neurogenic thoracic outlet syndrome

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Objective: **Cervical and anomalous first ribs are rare conditions, occurring in less than 1% of the population. This manuscript reviews our management of neurogenic thoracic outlet syndrome (TOS) associated with these congenital anomalies.**

Methods: **During the past 26 years, 65 operations were performed for abnormal ribs that produced symptoms of TOS. Of these, 54 operations were for neurogenic TOS and are covered in this paper. Indications for surgery were disabling pain and paresthesia and failure to respond to conservative treatment. Surgical technique for neurogenic TOS was supraclavicular cervical rib resection and scalenectomy without first rib resection in 22 cases, supraclavicular cervical and first rib resection in 17 cases, supraclavicular excision of anomalous first ribs in five cases, and transaxillary anomalous first rib resections in two cases (total, 46 cases). Eight reoperations were performed for recurrent TOS in patients who previously had undergone cervical and first rib resections.**

Results: **Neck trauma was the cause of neurogenic symptoms in 80% of patients with cervical or anomalous first ribs. The surgical failure rate was 28% for 46 primary operations. A significant variable in results was the etiology of the symptoms. The failure rate for patients in whom symptoms developed after work-related injuries or repetitive stress at work was 42%, and the failure rates for patients whose symptoms followed an auto accident or developed spontaneously were 26% and 18%, respectively. The failure rate in each etiology group also was affected by the operation performed. The failure rate for cervical rib resection without first rib resection in the work-related group was 75% compared with a failure rate of 38% in the non-work-related group. In contrast, when both cervical and first ribs were resected, the failure rate in the work-related group fell to 25% and in the non-work-related group to 20%. These failure rates for the work-related and non-work-related groups are similar to our failure rates in patients without cervical ribs.**

Conclusion: **Surgery for neurogenic TOS in patients with cervical ribs should include both cervical and first rib resection. The presence of cervical or anomalous first ribs in patients with neurogenic TOS does not improve the success rate from surgery compared with patients without abnormal ribs. Neck trauma is the most common cause for neurogenic TOS in patients with abnormal ribs. Cervical and anomalous first ribs are the predisposing factors rather than the cause. (J Vasc Surg 2002;36:51-6.)**

Cervical and anomalous first ribs are usually incidental findings on routine chest x-rays. Although most of these ribs produce no symptoms and necessitate no therapy, a few are symptomatic and do need treatment. In addition, certain asymptomatic cervical and anomalous first ribs present potential limb-threatening complications and should be followed at regular intervals.

During the past 28 years, we have performed 65 operations on patients with cervical or anomalous first ribs. Fifty-four operations were for neurogenic thoracic outlet syndrome (TOS), nine for complications of arterial TOS, and two for venous TOS. This manuscript will review our experiences with neurogenic TOS associated with cervical and anomalous first ribs.

MATERIAL AND METHODS

Among the more than 1000 operations performed for neurogenic TOS between 1972 and 2000, 42 patients had cervical or anomalous first ribs. Bilateral operations were performed in 12 patients, and unilateral procedures in 30. Thirty-nine cervical ribs were excised in 32 patients, seven anomalous first ribs were excised in five patients, and eight operations were performed on five patients who previously had undergone transaxillary first and cervical rib resection (except one who previously had a supraclavicular cervical rib resection) and now had recurrent neurogenic TOS. In the group that underwent unilateral operations, several patients had a cervical rib on the contralateral side that was not removed because it was asymptomatic. Thirty-five of the 39 patients in whom age was recorded were between 20 and 50 years of age; three were less than 20 years old, and one was more than 50 years old. Thirty-three patients were female (79%), and nine were male (21%).

Diagnostic studies. Radiograph of the chest and cervical spine, including the oblique view, was the primary study for diagnosis of cervical and anomalous first ribs (Figs 1 and 2). Electromyography/nerve conduction velocity studies were performed when diagnoses of carpal or cubital tunnel syndrome were suspected, but they were not performed routinely. Patients with long complete cervical or abnormal first ribs underwent screening for subclavian artery dilatation or aneurysm formation with duplex scan or arteriography.

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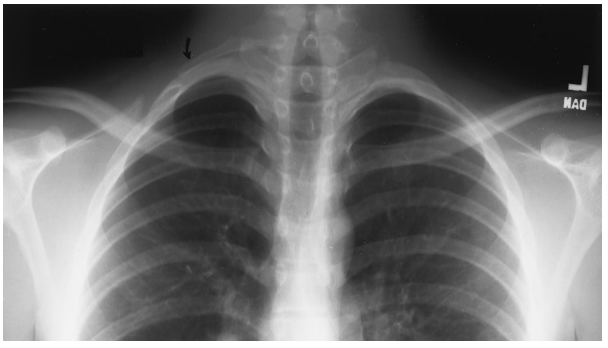

Fig 1. Complete right cervical rib *(arrow)* in 17-year-old swimmer with true joint between first and cervical ribs, excised through supraclavicular incision. Addition of infraclavicular incision at times can facilitate this excision, although it was not necessary in this patient.

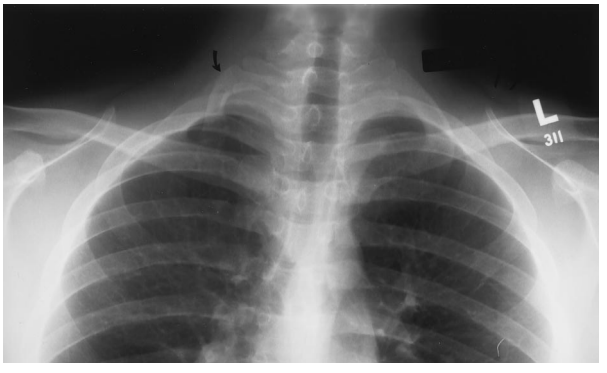

Fig 2. Anomalous right first rib *(arrow)* in 17-year-old male patient.

Indications for surgery. The primary surgical indications were failure to improve after a minimum of 3 to 12 months of conservative therapy and symptoms that were at least partially disabling. Patients with severe symptoms and complete cervical ribs underwent operation after shorter periods of conservative therapy. Conservative therapy consisted of several methods of physical therapy, including neck stretching exercises, abdominal breathing exercises, posture correction, nerve glides, and after 1998, the Feldenkrais method. Although we regard subclavian artery dilatation greater than 1.5 times the diameter of the proximal artery as an indication for rib resection even if the patient's neurogenic symptoms alone do not meet the criteria for surgery, no patients were seen in this category.

Surgical technique. The supraclavicular approach was used for all primary and secondary operations, except for three transaxillary procedures in 1974: two for anomalous first ribs and one reoperation in a patient with recurrence after supraclavicular cervical rib resection. The supraclavicular approach included both anterior and middle scalenectomy and the cervical rib resection in all cases. When the cervical rib was fused to the first rib with a true joint, the first rib also was removed to excise the joint and avoid leaving a rough or weakened first rib. However, in patients with short cervical ribs that joined the first rib via a ligament, first rib removal was optional. If the first rib could easily be removed, we usually did so; if its position was deep and wide, we left the first rib alone. Thus, first rib resection was not randomized.

In addition to anterior and middle scalenectomy, cervical and first rib resection was performed in 17 primary operations and cervical rib without first rib resection was performed in 22 operations; scalenectomy with excision of an anomalous first rib was carried out in five procedures; and scalenectomy with excision of stumps of first and cervical ribs was the operation for seven recurrences. The techniques of the supraclavicular and transaxillary approaches have previously been described.[1,2]

Table I. Etiology of symptoms

	No. of ribs
Neck trauma	43 (80%)
Auto accident	24 (44%)
Repetitive stress/work injury	17 (31%)
Other neck trauma	3 (5%)
Spontaneous	11 (20%)

Exception: complete cervical ribs had 50% spontaneous onset.

RESULTS

Etiology. The onset of symptoms was preceded by a history of neck trauma in 80%of the cases, and in 20% the symptoms developed spontaneously. However, in the subgroup of 12 patients with complete cervical ribs, six patients had the spontaneous onset of symptoms, for an incidence rate of 50% (Table I).

Symptoms. Symptoms included pain in the neck, trapezius, anterior chest wall, shoulder, arm, elbow, and forearm; paresthesia in the fingers, usually all five and often worse in the fourth and fifth fingers; occipital headaches; weakness in the hand and arm; aggravation of the symptoms with elevation of the arm; and coldness, color changes, and hand swelling. Although all symptoms were not present in each patient, most symptoms were. The incidence rate of each symptom was similar to that of patients with TOS without cervical ribs.[3]

Physical examination. Common findings were scalene muscle tenderness, a positive Tinel's sign over the brachial plexus, duplication of the symptoms with thumb pressure over the anterior scalene muscle, reproduction of symptoms with rotation of the neck to the contralateral side and tilting of the head to the contralateral side, and duplication of symptoms with the arms abducted to 90 degrees in external rotation (90-degree abduction in external rotation position). Three of the patients in this series had hand atrophy.

Rib anatomy. Cervical rib length was noted in 40 of the 47 operations for neurogenic TOS with cervical ribs. The ribs were 1 to 2 cm in 11 instances, 2 to 5 cm in 17

Table II. Types of ribs

	No. of ribs
Incomplete cervical rib	28 (59%)
Complete cervical rib	12 (26%)
Anomalous first rib	7 (15%)
Total ribs	47*

*In seven patients, rib length was not stated.

instances, and complete cervical ribs in 12 instances. Of the 12 complete ribs, three had a true joint at the first rib junction and the other nine had a fibrous attachment to the first rib (Table II).

In seven instances, anomalous first ribs were noted. They were positioned a little higher than the normal first rib, ran in a more vertical position, were thinner, and inserted into the lateral aspect of the second rib rather than the costal cartilage near the sternum. Initially, some of these had been diagnosed as cervical ribs. Identification as an anomalous first rib occurred with the realization of only one rib above the second rib with its origin appearing to be from the transverse process of thoracic vertebrae 1, rather than the seventh cervical vertebrae, although this was often hard to ascertain.

Complete versus incomplete cervical ribs. Incomplete cervical ribs acted differently than complete ribs. Incomplete ribs were associated only with neurogenic TOS, and complete cervical ribs produced neurogenic or arterial TOS. All arterial TOS cases as the result of cervical ribs in our experience, and in those of other investigators,[4] were associated with complete cervical ribs. Complete cervical ribs accounted for 18% of the neurogenic TOS cases in this study, incomplete cervical ribs for 67%, and abnormal first ribs for 15%. Seventy-five percent of patients with incomplete cervical ribs had the onset of their neurogenic symptoms after neck trauma, and only 50% of patients with complete cervical ribs had trauma as the cause. During the same time period, we saw nine patients with arterial TOS, four cases caused by complete cervical ribs and five by anomalous first ribs. Two other patients had venous TOS caused by an anomalous first rib.

Electrodiagnostic studies. In the three patients with hand atrophy, electromyography/nerve conduction velocity studies showed findings typical of ulnar neuropathy. In all other patients, these studies were usually normal but occasionally revealed nonspecific changes.

Surgical results. Results were classified as follows: excellent, relieving all symptoms; good, relieving most major symptoms, with minor symptoms persisting; fair, relieving some symptoms, but with most major symptoms persisting; or failure, indicating no significant improvement in any symptoms. Patients with conditions classified as fair stated that they had noticed enough improvement to have made the operation worthwhile.

The length of the follow-up period was less than 1 year in six patients (11%), 1 to 2 years in 10 patients (13%), and more than 2 years in 40 patients (76%), 10 of whom were followed beyond 5 years. From previous studies with life table methods, we learned that more than 80% of the failures occurred within the first 2 years.[3] Therefore, from a practical point of view, all follow-up studies beyond 2 years were grouped together.

The early results (first 3 months after surgery) of 46 primary operations were good to excellent in 33 (72%) cases, fair in eight cases (17%), and failure in five cases (11%). The late results were good to excellent in 27 cases (59%), fair in six cases (13%; total 72%), and failure in 13 cases (28%; Table III). Five of the 13 failures (13%) were immediate, three more occurred within the first year, four occurred between 1 and 2 years, and one occurred between 2 and 3 years.

Seven reoperations were performed on patients who previously underwent transaxillary first and cervical rib resections, and one reoperation was performed on a patient who previously had undergone a supraclavicular cervical rib resection. Reoperation with supraclavicular scalenectomy and resection of posterior rib stumps in seven patients and transaxillary first rib resection in one patient resulted in five failures in eight operations (63%). Three failures occurred within the first 9 months, and two between 1 and 2 years.

The following three clinical variables were compared: type of operation, etiology of symptoms, and complete versus incomplete cervical ribs. All differences noted were trends; none were statistically significant. Cervical rib resection with first rib resection had a lower failure rate (24%) than did cervical rib resection without first rib resection (41%; with χ^2 test, $P = .25$). Although the difference in failure rates was not statistically significant, the power to detect a difference of the observed magnitude was only 19%, given the small sample sizes as the result of low occurrence of TOS with cervical ribs.[5]

Work-related causes had a higher failure rate than did non-work-related accidents or spontaneous causes: 42% for 12 patients with work-related causes, 26% among 23 accident-related cases, and 18% among 11 patients with spontaneous onset (with Fisher exact test, $P = .29$). Within the work-related and non-work-related accident causes, the addition of first rib resection to cervical rib resection reduced the failure rate 75% to 25% (with Fisher exact test, $P = .22$) for work-related causes and 38% to 20% (with Fisher exact test, $P = .61$) for non-work-related accidents (Table III). Here again, although these differences were not statistically significant, insufficient power existed to adequately access the difference between the groups (15% power for work-related causes and 1% for non-work-related causes). Nevertheless, a trend favoring combined cervical and first rib resection was apparent. In the group of nine patients with spontaneous onset, no difference was found between removal of and no removal of the first rib.

Among the seven operations for anomalous first ribs, the cause of symptoms was an auto accident in five patients and spontaneous onset in two. No patients in this group had work-related causes. No failed operations were seen in this group.

Table III. Operations, etiologies, and failures

	Total operations		*Nonwork accident**		*Spontaneous onset†*		*Work accident or RSI*	
Operation	*No. of operations*	*No. failed*	*No. of operations*	*No. failed*	*No. of operations*	*No. failed*	*No. of operations*	*No. failed*
Cervical rib resection without first rib resection	22	9 (41%)	13	5 (38%)	5	1 (20%)	4	3 (75%)
Cervical rib with first rib resection	17	4 (24%)	5	1 (20%)	4	1 (25%)	8	2 (25%)
Anomalous first rib resection	7	0	5	0	2	0	0	0
Total primary operations	46	13 (28%)‡	23	6 (26%)	11	2 (18%)	12	5 (42%)
Reoperations§	8	5 (63%)	4	3 (75%)	4	2 (50%)	0	

Nonwork related (nonwork + spontaneous) operations, 34 patients; failed, 8 (25%); work-related operations, 12 patients; failed, 5 (42%).
*Nonwork accident includes auto accident and other neck injuries.
†Spontaneous onset, no history of neck trauma.
‡In 46 primary operations, results were 13 failures (28%), 6 fair results (13%), and 27 good to excellent (59%).
§Reoperations, after failure of transaxillary or supraclavicular rib resection.
RSI, Repetitive stress injury.

Patients with complete cervical ribs had better results than did those with incomplete cervical ribs. Of 12 operations for complete cervical ribs, only one failure (9%) was seen, with two fair results (16%) and nine good or excellent results (75%).

DISCUSSION

Although cervical ribs occur in less than 1% of the population, they were present in 4.5% of our patients with neurogenic TOS.[6] Other studies have reported an incidence rate of cervical ribs in patients with TOS as high as 57%.[6] The explanation for this large discrepancy probably lies in a difference in referral patterns. Some physicians are more reticent than others to refer patients with TOS symptoms for surgery unless an objective sign is seen, such as a cervical rib. However, in either case, this indicates that a person with a cervical rib is several times more likely to have TOS develop than a person without a cervical rib.

Cervical ribs are twice as common in women as men (68% versus 32%, respectively), are bilateral in more than 50% of cases, and are asymptomatic in 90% of cases.[3] The 10% of patients with cervical rib who are symptomatic usually have neurogenic symptoms, but some have arterial symptoms. In our experience, cervical ribs resulted in 39 operations for neurogenic TOS compared with four for arterial TOS. Anomalous first ribs produced seven operations for neurogenic TOS compared with five for arterial TOS.

Anomalous first ribs also are uncommon, with an incidence rate of 0.34% and a female:male ratio of 50:50.[3] We have recognized seven patients in whom an anomalous first rib was thought to contribute to neurogenic TOS, but there could have been other patients in whom an anomalous first rib was overlooked. Anomalous first ribs are harder to recognize than cervical ribs because they originate from the transverse process of the first thoracic vertebrae, just like normal first ribs, and are not recognizable on oblique neck x-rays, as are cervical ribs. Some anomalous first ribs lie higher than normal, giving the appearance of cervical ribs, and insert into the lateral portion of the second rib instead of into the sternum. These variations have not been classified, and they all fit under the general heading of "anomalous first ribs." Careful comparison with the rib anatomy of the contralateral side reveals an equal number of ribs on each side, with the rib above the second rib appearing smaller, thinner, and higher and fusing to the lateral portion of the second rib on the abnormal side.

In this review, neurogenic TOS developed in 34 patients with cervical ribs compared with seven patients with anomalous first ribs. This difference was not the same for arterial TOS in which, in our experience with nine cases, four were caused by cervical ribs and five were caused by anomalous first ribs. We have no good explanation for this. One can only theorize that cervical ribs irritate the brachial plexus much more often than the subclavian artery, whereas anomalous first ribs are more apt to compress the subclavian artery as often as the brachial plexus.

The pathophysiology of cervical ribs and anomalous first ribs is on the basis of their anatomic relationships, lying in a position to narrow the scalene triangle through which the brachial plexus and subclavian artery must pass. In a few patients, this narrowing, even when slight, is just enough to produce symptoms, but in the large majority of patients, it is not. Cervical and anomalous first ribs are predisposing factors for the development of TOS, but in most patients, more than an osseous abnormality is necessary to produce symptoms.

The cause of symptoms in most patients with cervical ribs was the same as in patients without cervical ribs. In 80% of our patients, a history of neck trauma preceded the onset of symptoms in patients with cervical ribs, an incidence rate that is almost identical to the incidence rate of trauma in patients with TOS without cervical ribs. Not only was the incidence rate of trauma similar in patients with or without cervical ribs, the postoperative failure rate was essentially the same: 28% failure in this study compared with 31% for patients with TOS without cervical ribs.[6]

The significance of this is seen with a comparison of two groups of patients with neurogenic TOS: those with and those without cervical ribs. Patients with cervical ribs certainly fulfill the criterion of showing an objective abnormality to support the diagnosis. Those without a cervical rib lack an objective sign. Yet, both groups have an 80% incidence rate of trauma as the cause of their symptoms, and both groups have similar failure rates of about 30%. This suggests that the presence of a cervical rib does not make the diagnosis of neurogenic more reliable and that the cause of surgical failure is most likely scar tissue in the healing process and seldom an error in diagnosis.

Indications for surgery. The presence of a cervical rib or anomalous first rib, by itself, is not an indication for surgery. Surgery is indicated only in the presence of symptoms that fail to respond to conservative therapy and produce disability. These are the same indications as for patients without rib abnormalities. Cervical ribs are objective findings, which all physicians seek in establishing a diagnosis. However, the surgical success rate for patients with cervical ribs is no better than the surgical success rate for patients with TOS without cervical ribs. This has been reported previously,[7] was confirmed in this study, and supports the view that a diagnosis of neurogenic TOS should be made primarily on the basis of clinical findings, with little reliance on the presence or absence of an osseous abnormality.

However, that complete cervical ribs and anomalous first ribs can cause arterial stenosis or aneurysms in the subclavian artery and that these can remain asymptomatic until thrombosis or embolization occurs should be pointed out. Therefore, when a complete cervical or anomalous first rib is found in a patient whose neurogenic symptoms do not meet the criteria for surgery, we evaluate and follow such a patient with duplex scanning for stenosis or aneurysm formation. We have been doing this for the past 4 or 5 years and, to date, have not found any arterial abnormalities. However, although no data support our practice, with the knowledge that serious arterial complications are associated only with complete cervical ribs and anomalous first ribs, we believe this prophylactic approach for following this small group of patients is reasonable.

Excision of the cervical rib with or without the first rib. Until we evaluated the patients in this study, no data existed to establish criteria for removal of the first rib at the time of cervical rib resection. The decision of whether or not to remove the first rib was an arbitrary one. However, our policy is now changing because the results of this study revealed a failure rate of 41% for cervical rib resection without first rib resection versus a failure rate of only 24% for combined cervical and first rib resection. This difference did not reach statistical significance probably because of the small number of patients, but it does present a trend that appears real. Other investigators also have noted failure to relieve symptoms when only the cervical rib was excised.[8] Currently, we to try to remove both first and cervical ribs whenever it is technically feasible and safe. No functional or structural disability appears to result from removal of both ribs.

Support for routine removal of the first rib is found in a comparison of the success rates in treatment of neurogenic TOS in patients without cervical ribs, where results indicated a 16% higher success rate in those patients with anterior and middle scalenectomy with first rib resection compared with those in whom the first rib was not removed.[9] This compares with the difference of 17% noted in this study for patients with cervical ribs between those who did and did not have the first rib excised.

The explanation for better results when the first rib is removed along with the cervical rib is not obvious. We theorize that the primary reason for surgical failure is brachial plexus compression by scar tissue formation in the first 18 months after surgery. If the first rib is present, the scar tissue can fix to the rib, whereas if the first rib is gone, scar tissue can fix only to the softer pleura.

Etiology of symptoms. Eighty percent of the patients had a history of neck trauma, and 20% had spontaneous onset. Neck trauma in the work place, either a single accident or repetitive stress injury, was followed by a higher failure rate than when the cause was non-work-related trauma, auto accidents, or spontaneous onset. In the work-related group, the failure rate for cervical and first rib resection was 42% compared with 26% for the same operation in the non-work-related group. The poorer results in patients whose onset of symptoms was related to neck trauma at work in patients with cervical ribs has also been observed in patients without cervical ribs.[9] We have no proven explanation for this difference between the work-related and non-work-related trauma groups.

Supraclavicular versus transaxillary approach. Either approach can successfully deal with cervical and anomalous first ribs. We prefer the supraclavicular approach because it provides better exposure of the posterior portion of both ribs and allows complete and safer excision of both first and cervical ribs back to the transverse processes. This approach also permits total anterior, middle, and minimus scalenectomy and excision of congenital bands that are often found lying over the eighth cervical and first thoracic nerve roots. In addition, it permits easier access to evaluate, open, and repair the subclavian artery when necessary. In a few patients, removal of a large prominence on the first rib is difficult through the supraclavicular incision alone. In such cases, the addition of an infraclavicular incision[8] or stretching of the supraclavicular incision below the clavicle[10] has been helpful.

CONCLUSION

The presence of a cervical rib, by itself, is not an indication for surgery in patients with neurogenic TOS. Other indications include failure to respond to conservative therapy and some degree of disability. Surgery should include resection of the first rib and the cervical rib. In neurogenic TOS, the surgical failure rate is similar for patients with or without cervical ribs. Neck trauma was the cause of neurologic symptoms in patients with cervical or

anomalous first ribs in 80% of patients, the same incidence rate as in patients without cervical or anomalous first ribs. The abnormal rib is a predisposing factor, not the cause. The failure rate for cervical rib resection, with or without first rib resection, is worse for patients whose symptoms are brought on by work-related neck trauma than in patients whose symptoms are the result of non-work-related neck trauma or a spontaneous onset. Complete cervical ribs act differently than incomplete cervical ribs. For neurogenic TOS, complete ribs have a 50% incidence rate of spontaneous onset of symptoms compared with 20% for incomplete ribs. Only complete ribs have been noted to produce arterial TOS.

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